CONGENITAL HEART DISEASE IN YAKIMA 2017

How to manage infants and children with congenital heart disease in central WA surrounded by mountain passes in winter and forest fires in summer
CENTRAL WA PEDIATRIC CARDIOLOGY SERVICES

- CLINICS 3/WEEK CHILDRENS VILLAGE YAKIMA 3/WEEK TRICITIES 1-2/WEEK WENATCHEE
- 40 pats/week  2000  per year  emergency add-on open spot every clinic

- ECHO 800-1000/yr  HOLTER/EVENT MONITOR  FETAL ECHO 10/month

- REFER  SCH FOR diagnostic/ interventional/EP CATH 65/yr and OHS 40-50 /yr

- OTHER SERV  in (chi vill) feeding/neurodev/fam services/orthoped/GI/GENETIC
  (tricities ) nephro/uro/gen surgery telemed derm
  (Wenatchee) ADCHD endocrine
SEATTLE CHILDRENS SERVICES

- OPEN HEART SURGERY  400-450 /yr  4 surgeons (mary bridge hospital Tacoma)
- Diagnostic/INTERVENTIONAL CARDIAC CATH  4 providers
  - PV/ ao valve/coarct balloon  PDA/ASD device  coA/ductal stent
- ELECTROPHYS/ABLATION
- TRANSPLANT/HEART FAILURE  3 providers
- PAH/CPET
- MRI CT  3D reconstruction for planning of OHS
- Genetics/22 q 11 clinic / MD clinic / vascular anomaly clinic
- ADCHD  3 providers
PEDIATRIC CARDIOLOGY AND CARDIAC DISEASE IN CHILDREN AND ADOLESCENTS

L-R SHUNTS:  VSD  ASD  PDA  AVSD (AVC /ECD)

R-L SHUNTS :  TOF  D-TGA  Tri ATRESIA  PA-VSD  PA-IVS  TRUNCUS  TAPVC  EBSTEINS

OBSTRUCTIVE LESIONS:  PS  CoA  HPLHS  AS (subAS supraAS )

POSTINFECTIOUS :  ARF/RHD  KAWASAKI  ENDOCARDITIS  PERICARDITIS

CONNECTIVE TISSUE DISEASE:  MARFANS  LOEYES-DIETZ  EHLER-DANLOS

CARDIOMYOPATHIES:  HCM/HOCM  DCM
COMMON GENETIC SYNDROMES ASSOCIATED WITH CHD
CHROMOSOMAL ABNORMALITIES 50-80% (1% IN FT)

DOWN'S: (CAVC TOF  DIGEORGE/VCF:  ( IAA TOF-PA/APV Truncus)  test hypocalcemia
WILLIAMS: ( SUAS PPS)  remember ?  vit D supplementation
ALAGILLE: (PPS)bil atresai
TURNER  BAV/COA  AAo anevr  NOONANS:  PS/HCM
CONGENITAL HEART DEFECTS.....

20% VSD

5-10% ASD PS PDA TOF HPLHS

3-6% CoA AS TGA

2-3% PA-IVS Tri A

1-2% PA-VSD DORV Truncus TAPVC

0.5-1% Ebsteins CCTGA SV APwindow Cor Triatriatum
**CLINICAL PEARLS**

SHUNT PRECORDIUM + INFLOW RUMBLE indicates QP/QS >2:1 and need for surgery

Shunt vascularity on CXR = vx>bronchiole

BP gradient – cuff on RA LA and one leg and rotate attmt – note HR

Large VSD can result in FTT without loud murmur as PVR falls
Some ASD can enlarge with body growth

LARGE PDA - rapid CHF since cor perfusion occurs in diastole-Aodias P decreased-LVEDP increased and time in dias shortened

FEM PULSES-decreased indicates CoA INCREASED/bounding in CHF = PDA/AP window or AR

LOUDER(holosys) murmur = higher Pgrad ie small defect or outflow obstruction-(ejection murmur)
CLINICAL PEARLS …

Cyanosis= R-L shunt   Recognition requires minimum of 3% desat Hb so delayed in anemia

TACHYPNEA with min cyanosis maybe DTGA - VSD or TAPVC

INFRADIAPHRAGMATIC TAPVC – obstructed gives small heart and “RDS” picture on CXR

VALVAR PS in infancy can progress from mild to severe or resolve in first 6 mo

BAV can develop first clinical sign in adulthood – endocarditis may be presenting symptom
40% of HPLHS have parent with BAV
HETEROTAXY = AMBIGUOUS SITUS WITH MIDLINE LIVER

COMPLEX MOSTLY CYANOTIC LESIONS  RT ISOMERISM (bilat rt lungs) ASPLENIA +PS/PA

LT ISOMERISM  bilat left sides )POLYSPLENIA and anomalous venous conn

Identify LA –receiving pulm veins  narrow base hookshaped atrial appendage

RA –suprahepatic IVC + coronary sinus  LIMBUS OF FOR OVALE  triangular broadbased AA

LEFT MAINSTEM BRONCHUS TAKE OFF

INCIDENCE CHD high in-  ISOLATED DEXTROCARDIA AV CONCORD and NRGV  “pivotal “

ISOLATED DEXTROCARDIA WITH L-TGA

NORMAL  D-LOOP  morphological RV is RT HANDED and anterior

L-LOOP  morphological LV is  RT HANDED and anterior
COMPLICATIONS/LONGTERM FOLLOW UP POSTOP CHD

• TOF: PVR – chronic PR and Rvdilation  VT AR (large ao root) small arm

• FONTAN: “single ventricle pathway” from HPLHS to Tri A to unbalanced AVC Gleen obstruction chronic hepatic congestion---cirrhosis  PLE pulm AVM

• MUSTARD/SENNING: PV/SVC obstruction AFibFlutter

• ARTERIAL SWITCH: Su PS (LeCompte maneuver) Su AS and cor ostial narrowing
KAWASAKI

- >5DAYS FEVER
- CONJUNCTIVITIS nonpurulent
- ORAL erythema cracked lips strawberry tongue
- CERVICAL node
- RASH urticarial to scarlet fever
- EXTREMITIES swollen hands/feet and digits palmar erythema
- ESR CRP anemia urethritis arthritis
- HYDROPS gallbladder
- 8 DAYS window to treat IVGG 2 gm/kg/12 hrs ASA hi vs low dose
LONGTERM FOLLOW UP KAWASAKI

- Echocardiography at dx or hospital dc then at 6-8 weeks with CBC/CRP not ESR
- if CRP normal and echo neg for CAA at 6-8 weeks stop ASA and rtn for echo at 1 yr
- If small/medium aneuvr or persistent cor dilation then follow on ASA yearly 2D no contact sports
- Large aneyr ASA plus Coumadin Q 3-6 mo 2D

- ALL PATIENT COR COUNSELING RE BMI/BP/SMOKING
- Reasonable to start statin "early" if indicated
MAJOR    CARDITIS CHOREA ARTHRITIS ERYTH MARG SUBCUT NODULES
MINOR    fever arthralgia increased PR increased ESR /CRP asp test
Support evidence ASLO/ANTIDNAse titers
2 major or 1 major and 2 minor
antiAcarbohydrate peak 1mo post nl by 2 yrs styts pos in chro MR
Valve ds resolves in 70-80% if secondary prevention is maintained

ARF JONES 1944
LA benz PCN Q 28 DAYS
Sufadiazone 1 gm daily
CHOREA jack in the box milkmaids grip labile mood “ ADHD” poor handwriting choreiform movements ST VITUS DANCE 3 MONTS POST INF
ENDOCARDITIS

- JANEWAY nontender eryth/hemorrh palms soles
- OSLER nodes tender red lumbs fingers/toes
- ROTH spots “fluffy” white eyeground spots
- +RF IN 50 %
- 70% in CHD/1/1280 hosp may occur in NN with normal hearts
- 80% STREP (viridans/alphahemo) or coag neg STAPH
- <10% GRAM NEG “HACEK “
- 5% NEG BLOODCX
- PCN GENT or CEF GENT if prosthetic valve VANCO GENT
PROPHYLAXIS GUIDELINES

HIGH RISK
1. PRIOR HX/OF ENDOCARDITIS …IVDA
2. PROSTHETIC VALVE /SHUNTS prosthetic material
3. UNREPAIRED CYANOTICS REPAIRED +post patch x6 mo +residual defect
4. TRANSPLANT with VALVE DS

MODERATE RISK
1. BAV  2. MVP  3. RHD SLE  4. HCM
5. UNOP PDA VSD AVSD

AMOX /CLINDA/AZITHRO
INTRAUTERINE CIRCULATION AND CHANGES AT BIRTH

• CIRCULATION IN PARALLEL - R-L ATRIAL AND DUCTAL SHUNT to fill left side and provide bypass of lungs
• Only 8% combined cardiac output over isthmus and peripheral PA’s – “coarctation watch” and physiological PPS murmur
• RISK FACTORS - TYPE 1 and 2 DIABETES PRIOR sib/parent with CHD
• Exposure lithium/anticonvulsants/antidepressants pesticides? Automechanic beautician
• At birth = separation from placenta/PGE + increase in arterial O2 leads to normal closure duct and FO----30-40% adults have PFO ? Chronic migraine – potential risk of paradox emboli
• Normal decrease in PVR over 6mo (delayed recognition large VSD /ALCAPA
DUCTAL DEPENDENT LESIONS

FOR SYSTEMIC OUTPUT

HPLHS
CRITICAL AS
TIGHT COARCTATION

FOR PULMONARY BLOOD FLOW

HPRHS  PA  Tri ATRESIA
TOF-PA  or severe PS

MIXING

DTGA
<table>
<thead>
<tr>
<th>Condition</th>
<th>Description</th>
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<tbody>
<tr>
<td><strong>SVT</strong></td>
<td>AVNRE</td>
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<tr>
<td></td>
<td>WPW ANTI/ORTHODROMIC</td>
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<td>AFIB/AFLUTTER</td>
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<td>NN AFLUTTER</td>
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<td>ATRIAL ECTOPIC TACHYCARDIA</td>
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<td><strong>VT</strong></td>
<td>WIDE COMPLEX – SLOWER RATE THAN SVT/WPW</td>
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<td>CATECHOLAMINE SENS VT – bidir VT lethal in 30-50% bbloq ICD</td>
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<tr>
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<td>SUSTAINED VT 50% NORMAL HEARTS</td>
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<tr>
<td></td>
<td>Repaired TOF HCM DCM ARVD</td>
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<tr>
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<td>LQTS Electrolytes OD BRUGADA (RBBB and ST elevation)</td>
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<td>IDIOPATHIC FASCICULAR LV T-reentry in LV = RBB + LAD or RBB + RAD (DDX SVT with RBB resp to verapamil)</td>
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<tr>
<td><strong>LQTS</strong></td>
<td>ROMANO-WARD autosomal dominant</td>
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<tr>
<td></td>
<td>JERVELL-LANGE NIELSEN autosomal recessive associated with congenital deafness</td>
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<td>Deafness/seizure d/o requires baseline ECG</td>
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<td>CONGENITAL CAVB may present as arrhythmia – frequent Pac’s</td>
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SUDDEN CARDIAC DEATH

• HCM 36%
• COR ARTERY ANOMALY – (LEFT from RT COR SINUS) 24%
• MARFANS MVP AO ANEURYSMS 6%
• CHD 5%
• DCM MYOCARDITIS ARVD 3%
• COR ARTERY DISEASE 2%
• COMMOTIO CORDIS <1%
HCM/HOCM

- INFANCY- IDM regress after 3mo NOONANS or GSD (infants + RVOTO)
- INCREASED MURMUR with sudden standing decrease with squatting
- Abnormal ECG may precede symptoms
- 80% abnormal intramural cor – myocardial bridge – inherent diastolic dysfxn
- Mortality rate 1% /yr adult 2%/yr in children
- Genetic marker known start screening early
  - if fam hx of SCD follow echo until 18 then Q5yrs
- >30 mm IVS prob needs AICD
- Trained athlete with LVH-stop all exercise x 8 weeks and reecho
MATERNAL SLE

• SCREEN FOR CONDXTN DS IN UTERO-

• Refer for fetal echo if mom has SLE/RA or known +SSA
• 18-28 WEEKS gestation follow mechanical PR interval

• Risk CAVB 1-8% if mom has SSA/SSB
• 15-18% if prior child with CAVB
• 60 % WILL REQUIRE PACEMAKER over lifetime
CARDIOVASCULAR RISK FACTORS

• OBESITY SEDENTARY LIFESTYLE
• PREDIABETES/INSULINRES ACANTHOSIS NIGRICANS
• NORMAL FAST TRIGLYCERIDES 40
• IDEAL TOTAL CHOL 150-160 LDL 90-100 HDL 50-55
• HYPERTENSION
• FAMILY HX/OF EARLY MI <55 Y/OLD
• KAWASAKI ARTERIAL SWITCH PA-IVS COR reimplantations TRANSPLANT
• SCREEN ALL >20 or earlier if risk factors
• TREAT if fails diet x1 yr + LDL>160 with fam hx + 10 y/old LDL >190 2 risk fct

• Congen RBBB and athletic heart ? Increased risk of AF
INDICATION FETAL ECHOCARDIOGRAPHY

- Prior child with CHD/Parent with CHD
- DM type 1 type 2
- Teratogenic exposure
- Known chromosomal fetal defect/ fetus with multiple anomalies
- Maternal SLE
ADCHD

INCREASING POPULATION more complex interventions – EP interventional cath

Need for management of coronary heart disease/other cardiovasc risk

Difficulties in employment increased risk of mental d/o

Need for management of pregnancy – fetal echocardiography MFM
ACHD IN THE UNITED STATES

• 2010  273,000 adult survivors projected to be at least 510,000 by 2050 incidence of CHD of .9 %  - mod to severe .6 %  - 2% if BAV /AS/AR . WORLDWIDE 3/million

Increasing number of survivors and more complex lesions to follow

impact of neurological sequelae after several OHS during first 2 years of life/employment opportunities

Complex arrhythmia-diff acces thru past surgeries-failing systemic ventricles..